Short-term results of duodenum-preserving pancreatic head resection for solid pseudopapillary neoplasm in children at the National Children's Hospital

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ABSTRACT

Objective: To evaluate the results of duodenum-preserving pancreatic head resection for treating solid pseudopapillary neoplasm (SPN) of the pancreas in children at the National Children's Hospital. **Methods:** A descriptive, retrospective study of 11 patients diagnosed with pancreatic SPN and treated surgically at the National Children's Hospital from May 2022 to May 2024. **Results:** The average age of diagnosis in children was 10.9 years (range: 8-15 years), predominantly affecting female patients (81.8%). The main clinical symptoms were epigastric and left hypochondrial pain (81.8%). The average tumor size was 51.3 mm (range: 2.6-10 cm). Computed tomography (CT) imaging revealed a predominantly mixed structure in 9 out of 11 patients (81.8%). All patients underwent duodenum-preserving pancreatic head resection, with no severe intraoperative or postoperative complications. **Conclusion:** SPN is a rare pancreatic tumor with low malignancy, minimal invasiveness, and low rates of metastasis and recurrence. Thus, surgery is a safe and effective treatment method for children, with a low complication rate.

Keywords: Children's solid pseudopapillary neoplasm, pancreas.

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INTRODUCTION

Solid pseudopapillary neoplasms (SPNs) of the pancreas are rare exocrine pancreatic tumors, accounting for about 1% of pancreatic tumors. The first description of an SPN was made by Gruber Frantz in 1927, with a detailed pathological description provided in 1959.

SPNs are most commonly found in women of reproductive age and are rare in children. However, due to advancements in medical technology, the detection rate of SPNs in children is increasing. Diagnosing SPNs can be challenging because the clinical symptoms are often non-specific, and standard blood tests are not useful in diagnosis. Preoperative diagnosis relies on CT or MRI imaging, with histopathology being the gold standard for confirmation. Surgery remains the primary treatment option for this condition, offering a long post-operative survival time.

Therefore, in this report, we aim to evaluate the surgical outcomes of treating solid pseudopapillary neoplasms of the pancreas in children at the National Children's Hospital.

METHODS

Patients

Eleven patients were diagnosed with solid pseudopapillary neoplasms (SPNs) of the

pancreatic head who underwent surgery at the National Children's Hospital between May 2022 and May 2024, with histopathological confirmation of SPNs.

The inclusion criteria were as follows: patients, regardless of sex and age. Patients with histopathological types not found in the pancreas, incomplete medical records, or missing histopathological results were excluded.

Methods

A descriptive case series was conducted. All patients with histopathological results confirming pancreatic SPNs from the Pathology Department were recruited. The medical record numbers of each patient from the medical record storage department were then obtained. The clinical and laboratory characteristics were also recorded.

The study indicators included sex, clinical classification, radiological and laboratory characteristics, and surgical characteristics.

RESULTS

Two boys and nine girls were included in the study. The median age was 10,9 (8-15) years. Nine patients had abdominal pain, one had loss of appetite, and one was diagnosed during a health check-up.

The average tumor size was 5.13 cm (ranging: 2.6 to 10 cm). The tumor structure on CT was primarily of mixed type (81.8%); most tumors showed uneven contrast enhancement due to necrosis and intratumoral hemorrhage. (Table 1)

Table 1. *Paraclinical features*

Paraclinical features			N	%
CT	Metastasis	Lymph node metastasis	1	9
		Other organ metastasis	0	0
	Tumor	Solid	2	18.2
		Cystic	0	0
		Mixed structure	9	81.8
	Contrast enhancement	Intense	0	0
		Mild	5	63.6
		Uneven	6	54.5
	Size	< 3cm	2	18.2
		3-10cm	5	63.6
		> 10cm	2	18.2
AFP	Normal		11	100
Serum glucose	Normal (3,3-5,5 mmol/L)		11	100
Serum P-Amylase	Normal (< 53 U/L)		11	100
Serum Lipase	Normal (7-39 U/L)		11	100

All the 11 patients underwent open surgery. Among them, seven patients experienced no intraoperative complications; three cases involved pancreatic duct injury, which was managed with a pancreaticojejunostomy, and one case involved a common bile duct injury, which was repaired during the surgery. (Table 2)

Regarding early complications**,** none of the children experienced bleeding, pancreatic fistula, bile leakage, residual abscess, or immediate post-operative death. Regarding late complications, one patient developed acute pancreatitis in the second week after surgery. All patients were assessed for postoperative pancreatic function, and none of them showed signs of endocrine pancreatic insufficiency. Of the nine patients assessed for exocrine pancreatic function, 3 (33.3%) patients. (Table 2)

Table 2. *Surgical Characteristics*

Surgical Characteristics		**N**	**%**
Duodenum-preserving pancreatic head resection	With pancreatico-enteric anastomosis	3	27.3
	Without pancreatico-enteric anastomosis	8	72.7
Intraoperative complications	Common Bile Duct Injury	1	9
	Pancreatic Duct Injury	3	27.3
Postoperative complications	Exocrine pancreatic insufficiency	3 (n = 9)	33.3
	Acute Pancreatitis	1	9

DISCUSSIONS

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare exocrine pancreatic tumor, accounting for about 1% of pancreatic tumors [1]. It was first identified by Gruber Frantz in 1927, and in 1959, the author detailed the pathology of this lesion.

SPN of the pancreas has been referred to by various names in literature, including Gruber Frantz tumor, papillary and solid tumor, cystic papillary tumor, solid-cystic tumor, papillary cystic and solid epithelial neoplasm, benign or malignant papillary neoplasm of the pancreas, and pancreatic acinar cell carcinoma. Some researchers even categorize it under pancreatic endocrine tumors.

The pathogenesis of SPN remains unclear. There are two main hypotheses: one suggests they originate from multipotent pancreatic cells, while the other suggests a derivation from female genital ridge cells [2]. Various reports indicate SPN primarily affects women of reproductive age and may have a genetic predisposition [3][4]. The incidence in children is lower than in adults, but with advancements in medical technology, the detection rate in children is increasing.

Patients often present with nonspecific clinical symptoms, which may include abdominal pain, nausea, vomiting, palpable abdominal mass, or abdominal distension due to tumor compression of adjacent organs. Maimaijiang et al. reported that 50% of pediatric patients with SPNs were asymptomatic, 38.89% presented with abdominal pain, 22.22% had a palpable abdominal mass, and 11.11% had symptoms following abdominal trauma [1].

Routine blood tests, including glucose, amylase, and lipase, are generally not helpful for diagnosis. Tumor markers are typically normal, and SPNs are rarely associated with endocrine syndromes [1]. In our study, most patients had normal biochemical blood markers and normal AFP levels.

Ultrasound typically shows heterogeneous hypoechoic masses, including solid, cystic, and calcified components [5]. Preoperative diagnosis mainly relies on CT or MRI imaging. On CT, SPNs often appear as large, well-circumscribed masses with heterogeneous density due to internal hemorrhage and necrosis. Differential diagnosis includes pancreatic pseudocyst, adenocarcinoma, mucinous cystadenoma, mucinous cystadenocarcinoma, serous cystadenoma, pancreatoblastoma, and hemangioma [6]. CT helps evaluate the tumor's relationship with surrounding structures, lymph node metastasis, and distant metastasis. MRI provides superior soft tissue resolution, which is advantageous for assessing the tumor's relationship with the bile duct and pancreatic duct [7]. Endoscopic ultrasound (EUS) with biopsy is a minimally invasive diagnostic procedure with relatively low complication rates. EUS has a sensitivity and specificity of 56%-71% and 45%-97%, respectively, for diagnosing SPNs [8][9]. There are few studies on the use of EUS for staging pancreatic cystic neoplasms.

Histopathologically, SPNs resemble pancreatic neuroendocrine tumors, but definitive diagnosis often requires immunohistochemical staining. Shen et al. found that P504S, TFE3, SOX-11, and progesterone receptors are valuable for differentiating SPNs from other pancreatic tumors [10]. Common markers used for diagnosing SPNs include Beta-catenin, CD10, Chromogranin, and Vimentin, with Beta-catenin (+) being sufficient for diagnosis [1].

Surgical resection remains the primary treatment for SPNs. Surgeons should aim for complete resection while preserving pancreatic function as much as possible. SPNs have low malignancy, limited invasiveness, low rates of metastasis and recurrence, and patients have a good prognosis even with metastasis or recurrence [11][12]. For tumors in the pancreatic head, less invasive surgeries such as pylorus-preserving pancreaticoduodenectomy or duodenum-preserving pancreatic head resection can be considered. Lymph node metastasis is rare in SPNs, and extensive lymph node dissection is usually unnecessary even when metastasis is present [1].

Among our 11 patients, one case had lymph node metastasis; the patient underwent resection of the primary tumor and metastatic lymph nodes without extensive lymph node dissection. After 14 months of follow-up, no recurrence or further metastasis was detected.

All patients in our study underwent open surgery for duodenum-preserving pancreatic head resection, with intraoperative complications in 4 out of 11 patients (pancreatic duct injury in 3 patients and common bile duct injury in 1 patient). No postoperative bleeding, bile leakage, pancreatic fistula, or residual abscesses were noted. According to Beger et al. [13], overall complication rates are similar between duodenum-preserving pancreatic head resection and conventional pancreaticoduodenectomy, but severe complications like pancreatic fistula and postoperative pancreatic function impairment are significantly reduced in the duodenum-preserving group.

Complete resection (R0) is considered the definitive treatment for SPNs. The role of chemotherapy in SPNs is unclear. Adjuvant chemotherapy is typically unnecessary after complete resection and is only indicated for distant metastasis or unresectable disease [1][7].

Overall, the prognosis for SPNs is favorable. Kato et al. [14] reported long-term survival

over 10 years for patients with unresectable tumors, likely due to the tumor's slow doubling time (765 days). Factors such as incomplete resection, large tumor size, intraoperative tumor rupture, and male gender are considered risk factors for recurrence [15][16]. Invasion, recurrence, and metastasis are not contraindications for surgery. Current reports indicate a 2% recurrence rate for SPNs post-surgery, which is significantly lower than previous reports (5%-15%) [3]. The average recurrence time is 41 months, with most recurrences occurring within the first 5 years, but 25% of recurrences occur after 5 years, underscoring the need for long-term management to detect local recurrence or distant metastasis [3].

Limitations of our study include a relatively short average follow-up period, a small sample size, and the need for longer follow-up to evaluate recurrence, metastasis, and pancreatic function impairment post-surgery.

CONCLUSIONS

Solid pseudopapillary neoplasm (SPN) is a rare tumor predominantly found in young women. Its clinical manifestations are nonspecific, often leading to misdiagnosis or missed diagnosis. SPNs of the pancreas have a low malignancy potential, limited invasiveness, and low rates of metastasis and recurrence. Therefore, surgical intervention is a safe and effective treatment method for children, with a low complication rate.

REFERENCES

1. Maimaijiang, A., Wang, H., Li, W., Wang, Y.: Diagnosis and treatment of solid pseudopapillary neoplasm of the pancreas in children: A report of 18 cases. Front. Pediatr. 10, 899965 (2022). https://doi.org/10.3389/fped.2022.899965
2. Naar, L., Spanomichou, D.-A., Mastoraki, A., Smyrniotis, V., Arkadopoulos, N.: Solid Pseudopapillary Neoplasms of the Pancreas: A Surgical and Genetic Enigma. World J. Surg. 41, 1 (2017). https://doi.org/10.1007/s00268-017-3921-y
3. Yepuri, N., Naous, R., Meier, A.H., Cooney, R.N., Kittur, D., Are, C., Jain, A., Dhir, M.: A systematic review and meta-analysis of predictors of recurrence in patients with Solid Pseudopapillary Tumors of the Pancreas. HPB. 22, 12–19 (2020). https://doi.org/10.1016/j.hpb.2019.06.005
4. Wu, J., Mao, Y., Jiang, Y., Song, Y., Yu, P., Sun, S., Li, S.: Sex differences in solid pseudopapillary neoplasm of the pancreas: A population-based study. Cancer Med. 9, 6030–6041 (2020). https://doi.org/10.1002/cam4.3180
5. Buetow, P.C., Buck, J.L., Pantongrag-Brown, L., Beck, K.G., Ros, P.R., Adair, C.F.: Solid and papillary epithelial neoplasm of the pancreas: imaging-pathologic correlation on 56 cases. Radiology. 199, 707–711 (1996). https://doi.org/10.1148/radiology.199.3.8637992
6. Bochis, O.V., Bota, M., Mihut, E., Buiga, R., Hazbei, D.S., Irimie, A.: Solid pseudopapillary tumor of the pancreas: clinical-pathological features and management of 13 cases. Clujul Med. 1957. 90, 171–178 (2017). https://doi.org/10.15386/cjmed-672
7. Kumar, N.A.N., Bhandare, M.S., Chaudhari, V., Sasi, S.P., Shrikhande, S.V.: Analysis of 50 cases of solid pseudopapillary tumor of pancreas: Aggressive surgical resection provides excellent outcomes. Eur. J. Surg. Oncol. J. Eur. Soc. Surg. Oncol. Br. Assoc. Surg. Oncol. 45, 187–191 (2019). https://doi.org/10.1016/j.ejso.2018.08.027
8. Brugge, W.R., Lewandrowski, K., Lee-Lewandrowski, E., Centeno, B.A., Szydlo, T., Regan, S., del Castillo, C.F., Warshaw, A.L.: Diagnosis of pancreatic cystic neoplasms: a report of the cooperative pancreatic cyst study.

Gastroenterology. 126, 1330–1336 (2004). https://doi.org/10.1053/j.gastro.2004.02.013
9. Ahmad, N.A., Kochman, M.L., Brensinger, C., Brugge, W.R., Faigel, D.O., Gress, F.G., Kimmey, M.B., Nickl, N.J., Savides, T.J., Wallace, M.B., Wiersema, M.J., Ginsberg, G.G.: Interobserver agreement among endosonographers for the diagnosis of neoplastic versus non-neoplastic pancreatic cystic lesions. Gastrointest. Endosc. 58, 59–64 (2003). https://doi.org/10.1067/mge.2003.298
10. Shen, Y., Wang, Z., Zhu, J., Chen, Y., Gu, W., Liu, Q.: α-Methylacyl-CoA racemase (P504S) is a useful marker for the differential diagnosis of solid pseudopapillary neoplasm of the pancreas. Ann. Diagn. Pathol. 18, 146–150 (2014). https://doi.org/10.1016/j.anndiagpath.2014.02.006
11. You, L., Yang, F., Fu, D.-L.: Prediction of malignancy and adverse outcome of solid pseudopapillary tumor of the pancreas. World J. Gastrointest. Oncol. 10, 184–193 (2018). https://doi.org/10.4251/wjgo.v10.i7.184
12. Lubezky, N., Papoulas, M., Lessing, Y., Gitstein, G., Brazowski, E., Nachmany, I., Lahat, G., Goykhman, Y., Ben-Yehuda, A., Nakache, R., Klausner, J.M.: Solid pseudopapillary neoplasm of the pancreas: Management and long-term outcome. Eur. J. Surg. Oncol. J. Eur. Soc. Surg. Oncol. Br. Assoc. Surg. Oncol. 43, 1056–1060 (2017). https://doi.org/10.1016/j.ejso.2017.02.001
13. Beger, H.G., Nakao, A., Mayer, B., Poch, B.: Duodenum-preserving total and partial pancreatic head resection for benign tumors – Systematic review and meta-analysis. Pancreatology. 15, 167–178 (2015). https://doi.org/10.1016/j.pan.2015.01.009
14. Kato, T., Egawa, N., Kamisawa, T., Tu, Y., Sanaka, M., Sakaki, N., Okamoto, A., Bando, N., Funata, N., Isoyama, T.: A case of solid pseudopapillary neoplasm of the pancreas and tumor doubling time. Pancreatology. 2, 495–498 (2002). https://doi.org/10.1159/000064711
15. Del Chiaro, M., Verbeke, C., Salvia, R., Klöppel, G., Werner, J., McKay, C., Friess, H., Manfredi, R., Van Cutsem, E., Löhr, M., Segersvärd, R., Abakken, L., Adham, M., Albin, N., Andren-Sandberg, Å., Arnelo, U., Bruno, M., Cahen, D., Cappelli, C., Costamagna, G., Del Chiaro, M., Delle Fave, G., Esposito, I., Falconi, M., Friess, H., Ghaneh, P., Gladhaug, I., Haas, S., Hauge, T., Izbicki, J., Klöppel, G., Lerch, M., Lundell, L., Lüttges, J., Löhr, M., Manfredi, R., Mayerle, J., McKay, C., Oppong, K., Pukitis, A., Rangelova, E., Rosch, T., Salvia, R., Schulick, R., Segersvärd, R., Sufferlein, T., Van Cutsem, E., Van der Merwe, S., Verbeke, C., Werner, J., Zamboni, G.: European experts consensus statement on cystic tumours of the pancreas. Dig. Liver Dis. 45, 703–711 (2013). https://doi.org/10.1016/j.dld.2013.01.010
16. Lin, M.Y.C., Stabile, B.E.: Solid Pseudopapillary Neoplasm of the Pancreas: A Rare and Atypically Aggressive Disease among Male Patients. Am. Surg. 76, 1075–1078 (2010). https://doi.org/10.1177/000313481007601011